IN THE CLAIMS:

Please cancel claims 2-17 without prejudice, and amend claim 1 and add new claims 20-33 as follows:

sub c' 1. (Amended) A transgenic mouse model whose genome [contains] comprises [at least one] a mutation [that substantially reduces] capable of causing a reduced expression of [the] a native mouse *Smn* gene and [carries at least one] a human genomic DNA sequence [that at least partially compensates] capable of partially phenotypically compensating for the reduced expression of said mouse *Smn* gene.

sub c² 20. The transgenic mouse model of claim 1, wherein said mutation in said *Smn* gene is a knockout mutation and said human genomic DNA sequence contains a copy of human *SMN*^{c} gene.

21. The transgenic mouse model of claim 20, wherein said knockout mutation comprises an insertion in said mouse *Smn* gene of a hypoxanthine phosphoribosyl-transerase cassette.

22. The transgenic mouse model of claim 21, wherein said hypoxanthine phosphoribosyl-transerase cassette is inserted in exon 7 of said mouse *Smn* gene.

23. The transgenic mouse model of claim 20, wherein said knockout mutation comprises a replacement of exon 7 of said mouse *Smn* gene by a hypoxanthine phosphoribosyl-transerase cassette.

24. A method of generating a transgenic mouse model of spinal muscular atrophy, comprising the steps of:

(a) introducing a mutation in the genome of a mouse where said mutation causes a reduction of the expression of a mouse *Smn* gene to such a degree that said mouse would not survive; and

(b) introducing a human genomic DNA sequence comprising a copy of human *SMN*^{c} gene into the genome of said mouse where said human *SMN*^{c} gene expresses its itself to rescue said mouse.

25. The method of claim 24, wherein said mutation is a knockout mutation.

26. The method of claim 25, wherein said knockout mutation is introduced by inserting in said mouse *Smn* a hypoxanthine phosphoribosyl-transerase cassette or by replacing exon 7 of said mouse *Smn* with a hypoxanthine phosphoribosyl-transerase cassette.

27. The method of claim 26, wherein said human genomic DNA sequence further comprises a copy of centromeric *SERF1* and a portion of centromeric *NAIP*.

28. A method of testing for therapeutic efficacy on one or more symptoms of spinal muscular atrophy, said method comprising:

(a) applying one or more of therapies to be tested to a transgenic mouse model of claim 20; and

(b) determining whether one or more symptoms of spinal muscular atrophy have changed as a result of application of said therapy or therapies.

29. The method of claim 28, wherein said therapy is a gene therapy which corrects genetic defects by changing one or more genomic DNA sequences of said transgenic mouse model.

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30. The method of claim 28, wherein said therapy is a drug therapy which alleviates one or more symptoms of spinal muscular atrophy by using one or more chemical compounds.

31. The method of claim 28, wherein said transgenic mouse model is made according to claim 23.

32. The method of claim 31, wherein said therapy is gene therapy which corrects genetic defects by changing one or more genomic DNA sequences of said transgenic mouse model.

33. The method of claim 31, wherein said therapy is drug therapy which alleviates one or more symptoms of spinal muscular atrophy by using one or more chemical compounds.